pulmonary fibrosis patient information guide

[ENGLISH]

Pulmonary Fibrosis
FOUNDATION

contents

The mission of the Pulmonary Fibrosis Foundation (PFF)

is to help find a cure for idiopathic pulmonary fibrosis, advocate for the pulmonary fibrosis community, promote disease awareness, and provide a compassionate environment for patients and their families.

This educational guide is provided by the PFF as a public service to our patient community.

As a 501(c)(3) public benefit organization, the PFF provides information to the national and international pulmonary fibrosis community free of charge; we rely on public support to provide these valuable resources. If you value our work, please consider making a gift to the PFF. Your gift enables us to be a source of compassionate support for patients and their families and helps fund research to find a cure.

about this guide

PATIENTS DIAGNOSED WITH PULMONARY FIBROSIS (PF) OR IDIOPATHIC PULMONARY FIBROSIS (IPF), and their family members, often feel confused, concerned, and overwhelmed. Patients may find themselves frustrated by the lack of available information. Physicians often don't have the necessary time or resources to explain the details of the disease to their patients or to help patients deal with the trauma of being told that they have an incurable illness.

The Pulmonary Fibrosis Foundation is deeply aware of these concerns. We strive to provide patients, family members, and health care providers with the resources necessary to more fully understand PF and IPF, and to provide patients with the tools necessary to live with their disease and improve their quality of life.

It is important to note that care for PF or IPF is individualized, and disease progression varies greatly in patients—your physician may have discussed this challenge with you. Therefore, it is important for patients to understand their condition and ask their physicians important questions to ensure they are being treated appropriately based on their individual symptoms. This guide is intended to help patients achieve this goal.

Please remember that this information is a brief overview and is for educational purposes only. It is not intended to be a substitute for professional medical advice. Always consult your personal physician or health care provider with any questions you may have regarding your specific medical condition.

Also please know that we are here to help you. You may contact the Pulmonary Fibrosis Foundation with any questions or concerns you have about PF during the course of your care. Our staff can be reached at 888.733.6741 or by email at info@pulmonaryfibrosis.org.

defining pulmonary fibrosis

PULMONARY FIBROSIS (PF) DESCRIBES A CONDITION in which the lung tissue becomes thickened, stiff, and scarred.[1] The medical terminology used to describe this scar tissue is fibrosis. The alveoli (air sacs) and the blood vessels within the lungs are responsible for delivering oxygen to the body, including the brain, heart, and other organs. All of the body's functions depend upon delivery of a steady supply of oxygen. As lung tissue becomes scarred and thicker, it is more difficult for the lungs to transfer oxygen into the bloodstream. As a result, the brain, heart, and other organs do not get the oxygen they need to function properly.[1] In some cases, doctors can determine the cause of the fibrosis (scarring), but in many cases the cause remains unknown. When there is no known cause for the development of pulmonary fibrosis (and certain radiographic and/or pathologic criteria are met), the disease is called idiopathic pulmonary fibrosis or IPF. More specifically, consensus treatment guidelines from international lung societies define IPF as "a specific form of chronic, progressive fibrosing interstitial pneumonia of unknown cause, occurring primarily in older adults, limited to the lungs, and associated with the histopathologic and/or radiologic pattern of UIP [usual interstitial pneumonia]."[2]

There are more than 200 related diseases of the lung known as interstitial lung diseases (ILD), which are also referred to as diffuse parenchymal lung diseases or DPLD. Because these diseases affect the interstitium—the space around the alveoli—ILDs are classified as a group. However, ILDs may also affect other parts of the lungs. Many ILDs have similar characteristics to IPF and most result in lung fibrosis.

There is a subgroup of ILDs called idiopathic interstitial pneumonias (IIP), where the lung tissue becomes inflamed and scarring can also occur. The word pneumonia is used to describe inflammation and not an infection such as bacterial pneumonia. IIP is further broken down into a number of pathological subtypes. These subtypes include usual interstitial pneumonia (UIP), non-specific interstitial pneumonia (NSIP), desquamative interstitial pneumonia (DIP), respiratory bronchiolitis-associated interstitial lung disease (RB-ILD), acute interstitial pneumonia (AIP), cryptogenic organizing pneumonia (COP), and lymphocytic interstitial pneumonia (LIP).[2] IPF is a subtype of IIP, the pathological pattern seen in IPF is UIP. It is important for your care providers to differentiate the specific subtype of interstitial disease, since treatment, management, and prognosis can vary quite dramatically.

If there is a clear association with another illness or the lung scarring (fibrosis) is the result of a side effect from a medication or an exposure to an agent known to cause PF, then the cause of the disease is no longer considered idiopathic. PF clearly associated with another disease, such as scleroderma or rheumatoid arthritis, would be referred to as *pulmonary fibrosis secondary to scleroderma* or *secondary to rheumatoid arthritis*. As we learn more about interstitial lung diseases, the terminology may evolve.

prevalence and incidence

THERE ARE NO RELIABLE DATA TO DETERMINE how many people are affected by PF, possibly due to the large number of conditions under which it can arise. However, one recent study estimates the prevalence of all ILDs in the United States (US) at about 500,000, with IPF being the most common.[3] In the US, IPF affects between 132,000–200,000 people.[4] Approximately 50,000 new cases are diagnosed each year and as many as 40,000 Americans die from IPF each year.[4] There is limited information on the prevalence of IPF in the European Union (EU). The current estimate of the incidence of IPF in the EU is between 37,000 and 40,000 people, and in the United Kingdom more than 5,000 new cases are diagnosed each year.[5] More importantly, it is anticipated that the number of individuals diagnosed with IPF will continue to increase. This is likely to be a result of people living longer, an improved clinical understanding of IPF, and earlier and more accurate diagnosis.[6]

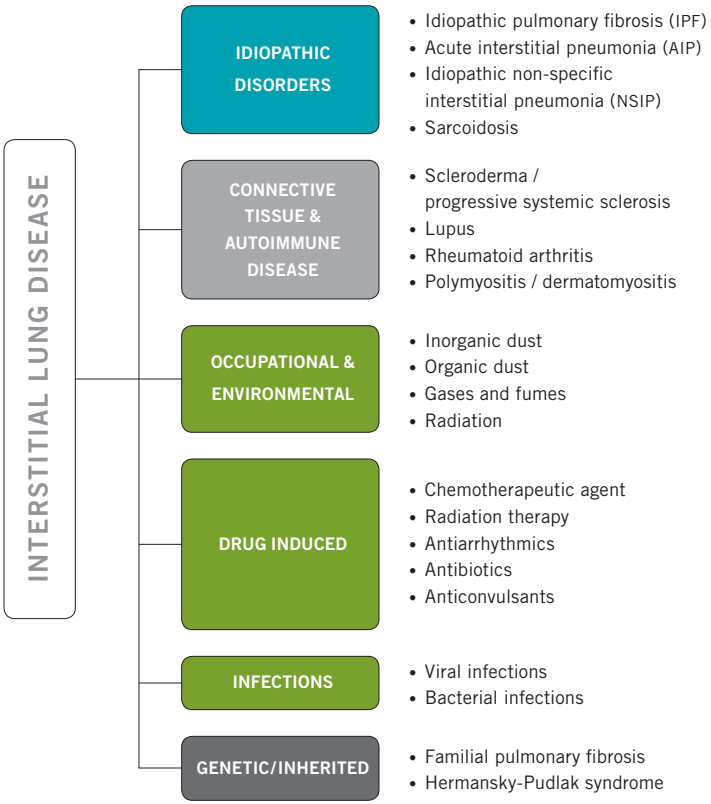
IPF has no specific demographic profile; it is found in equal proportions in urban and rural environments. A history of smoking and certain genetic factors has been associated with an increased risk of IPF, and a variety of published studies have indicated that about two-thirds of individuals with IPF have a history of smoking (see *Causes*).[7] IPF affects more men than women and usually occurs between the ages of 50 and 70.

The median age at time of diagnosis is approximately 63 years old according to a variety of published studies; however, IPF has been diagnosed in people from early adulthood into their late eighties.[6] It is clear that age is a significant risk for the development of pulmonary fibrosis.[6]

causes

PF CAN RESULT FROM A NUMBER OF CAUSES. As shown in the following figure, there are four general categories that can lead to the development of fibrotic lung disease: those of unknown or undetermined cause (teal), those that arise from a systemic autoimmune disease (gray), those that are associated with exposure to an agent known to cause PF (green), and those that have an inherited or genetic component (dark gray).

CLASSIFICATION OF COMMON INTERSTITIAL LUNG DISEASES



Systemic autoimmune diseases comprise a group of immunologic disorders that can result in the development of an IIP. These disorders include connective tissue diseases (CTD) such as rheumatoid arthritis (RA), lupus erythematosus (lupus), scleroderma (SSc), polymyositis/dermatomyositis, systemic vasculitis such as ANCA (+) polyangiitis (also called Wegener's granulomatosis), and Churg-Strauss syndrome. The specific type of lung disease that these disorders cause is based on the underlying autoimmune disorder.[8] The most common systemic autoimmune diseases that can result in the development of PF are SSc, RA, and lupus. Patients with CTD are also more likely to develop pulmonary hypertension (high blood pressure in the lungs). It is highly recommended that patients with CTD and symptoms of lung disease be evaluated and followed by a rheumatologist for their CTD and a pulmonologist for their lung disease.

PF can develop through **significant exposure to environmental or occupational agents**. Exposure to inorganic dust (asbestos, silica, beryllium, hard metal dusts), organic dust (animal proteins, bacteria, molds, fungi), or gases and fumes can contribute to the development of PF. Some of the most common forms of occupational related PF are asbestosis and silicosis. People that work in places or spend extended periods of time where there are high levels of organic dusts can develop hypersensitivity pneumonitis (HP). The two most common types of disease caused by organic dust are "bird fancier's lung" and "farmer's lung." Additionally, **exposure to some medications, high-dose radiation, and radiation therapy** can also result in the development of PF. Medications such as antibiotics (nitrofurantoin, sulfasalazine), antiarrhythmics (amiodarone, propranolol), anticonvulsants (phenytoin), and chemotherapeutic agents (methotrexate, bleomycin, oxaliplatin) have also been associated with the development of PF.

We know a lot about how some ILDs affect patients, but frequently a cause has not been identified. As its name suggests, the origin and development of **IIPs and other idiopathic disorders**, such as sarcoidosis, are still not completely understood. *The most common form of IIP is IPF.* The current thinking is that there is an abnormal response to injury that ultimately results in scarring of the lung. It is also hypothesized that certain environmental and genetic factors may contribute to the development of IPF; as these are more clearly defined, the disease process should be better understood. Ultimately, this should lead to new and effective treatments.

It is estimated that approximately 10–15% of patients with IPF have a form of PF that occurs in families. This is called **familial pulmonary fibrosis (FPF) or familial interstitial pneumonitis (FIP)**. In some families with FPF/FIP, not every affected family member has the same type of IIP. In about half of families with FPF, one or more family members have IPF and another has a different form of IIP. Other forms may include NSIP, DIP, RB-ILD, and COP.

Another rare form of familial or genetic PF is Hermansky-Pudlak syndrome (HPS). There are eight different types of HPS that can be distinguished by the signs, symptoms, and underlying genetic cause; types 1, 2, and 4 are the types associated with the development of PF.

As is suggested by FPF/FIP, there is a growing body of evidence suggesting that genes or genetic variants may predispose certain individuals to developing PF or IPF.

- Studies have found that some families with a history of more than two cases of IPF carry a mutation in the surfactant protein C (SP-C) gene, which normally helps lungs function correctly.[9]

- Another study suggested that the presence of specific genes may predict which IPF patients will have a more severe, rapidly progressing form of the disease.[10]
- Shortened telomeres (telomeres protect the fragile ends of chromosomes from deterioration) may be the cause of PF in certain patients as they grow older. Mutations in the genes *TERT* and *TERC* result in shortened telomeres and appear to predispose certain individuals to PF.[11,12]
- Individuals with variations in the *MUC5B* gene, which encodes a mucus protein, may have an increased risk between 6–22 times of developing PF depending on family history.[13]

The clinical implications of the identification of genetic variations that are associated with PF remain unclear, as there are no therapies targeting specific genetic factors for PF. Further, there is limited availability of genetic testing to identify genes that may contribute to PF or IPF. It is therefore important for patients to discuss the potential utility and possible risks of genetic testing with a qualified genetic counselor and their health care provider.

FACTORS ASSOCIATED WITH THE DEVELOPMENT OF PF

- Cigarette smoking
- Prolonged exposure to occupational or environmental contaminants or dusts
- Viral or bacterial lung infections
- Certain medicines, such as some antibiotics, antiarrhythmics, anticonvulsants, chemotherapeutic agents, or therapeutic radiation
- Acid reflux disease (GERD)
- Genetic predisposition

symptoms

SYMPTOMS MAY NOT BE PRESENT early in the disease and may not occur until the disease has progressed. Most patients will have a gradual worsening of lung function over time, although some will remain stable. Some patients may experience episodes of acute worsening of lung function without a clinically apparent infection or other cause; these episodes of acute worsening are called "acute exacerbations."

The most common symptom is shortness of breath, also known as dyspnea, which many patients describe as a feeling of breathlessness. Some patients, especially older patients, often ignore the occasional difficulty with breathing, attributing it to getting older or being out of shape. As the condition progresses and the damage to the lungs becomes more severe, breathlessness may occur with minor physical activity such as showering and getting dressed. Speaking on the phone and eating may also cause breathlessness with advanced disease. About 50% of patients with IPF may have "clubbing" of the fingertips due to a lack of oxygen in the blood. Clubbing is a thickening of the flesh under the fingernails, causing the nails to curve downward. Clubbing of the fingertips is not specific to IPF and occurs in other lung disorders, heart and liver disease, and can also be present at birth.

Other common symptoms include:

- Chronic dry, hacking cough
- Fatigue and weakness
- Discomfort in the chest
- Loss of appetite
- Unexplained weight loss

diagnostic tests and assessments

WITHIN THE GENERAL MEDICAL COMMUNITY the lack of clinical knowledge of PF remains a concern. This is further complicated by the fact that there are more than 200 different types of ILD. Limited awareness of the causes and an inadequate understanding of disease progression have resulted in misdiagnosis of many of the ILDs. In fact, a recent study showed that more than 50% of patients with IPF might have been initially misdiagnosed.[7]

It was not until 1999 that the American Thoracic Society (ATS) and the European Respiratory Society (ERS), in collaboration with the American College of Chest Physicians (ACCP), described specific clinical and pathological characteristics of IPF.[14] In 2011 the ATS, ERS, Japanese Respiratory Society (JRS), and the Latin American Thoracic Society (ALAT) defined evidence-based guidelines for the diagnosis and management of IPF, helping to standardize IPF diagnosis and treatment.[2] In general, the diagnosis of IPF requires three factors:[14]

1. Exclusion of other known causes of ILD.
2. The presence of a UIP pattern on high-resolution computed tomography (HRCT) in patients not subjected to surgical lung biopsy.
3. Specific combinations of HRCT and surgical lung biopsy pattern in patients subjected to surgical lung biopsy.

Your care provider will use a series of diagnostic tests and assessments to help determine if you have PF. To determine if you have PF, and the type of PF, you may require multiple tests. Additionally, the exclusion of known causes of fibrotic lung disease requires a careful

history and physical examination focusing on comorbidities, medication use, environmental exposures, and family history.

Multiple care providers from a variety of specialties, including pulmonologists, radiologists, rheumatologists, and pathologists, can help determine your diagnosis. Providers who are experienced in the diagnosis of PF can improve the accuracy of your diagnosis. To find a medical center near you with specific expertise in PF call 888.733.6741 or visit www.pulmonaryfibrosis.org/medicalcenters.

The tests outlined below may be used in the diagnosis of PF. If you have already been diagnosed with PF, many of these tests will be used to follow your lung health.

DIAGNOSTIC TESTS AND ASSESSMENTS

History and Physical Exam: A detailed medical history and examination to learn if there were any environmental, occupational, familial, or other medical conditions that could have contributed or predisposed a person to the disease's development. When listening to the lungs with a stethoscope, the physician may hear "crackles" or Velcro-like sounds. These are "opening" sounds made by the small airways during inspiration.

Chest X-Ray: A routine chest X-ray may be used as a screening test. However, 5–15% of patients with significant scarring will have a normal chest X-ray and IPF cannot be diagnosed from a chest X-ray alone.

High-Resolution Computerized Tomography (HRCT): A detailed image of the lungs to help physicians more clearly identify certain radiographic patterns in the lung tissue that may indicate disease. A radiologist may identify a "honeycombing" pattern that suggests lung scarring and damage to the air sacs, or "ground-glass opacity" that refers to the hazy appearance of lung tissue that is most associated with inflammation.

Pulmonary Function Tests: Breathing tests that measure the total amount of air in the lungs and assess the flow of air in and out of the lungs. There are two important components to pulmonary function tests: (1) spirometry that measures inspired and expired lung volumes and the rate at which this occurs and (2) diffusion capacity, or DLCO, that measures the ability of oxygen to diffuse into the bloodstream. These tests are usually done in a hospital or clinical laboratory and consist of breathing into a spirometer; they are sometimes done sitting inside a large plastic enclosure that resembles a glass telephone booth.

Pulse Oximeter: A device placed on the finger or earlobe that indicates the amount of oxygen saturation in the blood. Normal ranges are 95–100% on room air. Pulse oximetry does not measure carbon dioxide levels, so a blood gas level measurement may be necessary in some patients.

Arterial Blood Gas (ABG): Direct arterial puncture to measure arterial pH, oxygen saturation (PaO2), and carbon dioxide content (PCO2). Arterial blood has been oxygenated by the lungs and thus indicates how much oxygen is available to the body.

Bronchoscopy: Examination of the main airways of the lungs through the use of a small, flexible tube called a bronchoscope. Bronchoscopy helps to evaluate lung problems or blockages and provides a means to sample tissue or fluids. Unfortunately, the lung tissue samples obtained through bronchoscopy are small and usually inadequate for a definitive diagnosis.

Bronchoalveolar Lavage (BAL): A way to remove a tiny sampling of cells from the lower respiratory tract using a bronchoscope. A small amount of saline is injected through the bronchoscope and, when withdrawn, removes a sample of cells from the respiratory tract. Usually this is not helpful in making the diagnosis of IPF, but may be beneficial in other clinical situations.

Surgical Lung Biopsy: Surgical lung biopsy can be an important diagnostic tool in the evaluation of patients suspected of having a fibrotic lung disorder and is generally considered the "gold standard" for diagnosis. A lung biopsy in conjunction with an HRCT can also help determine how far the disease has progressed. Usually the biopsy can be performed by a minimally invasive procedure using video assisted thoracoscopic surgery (VATS). VATS is usually well tolerated, but it may not be recommended for all individuals.

Exercise Testing: A measure of how well the lungs function during exertion. Methods vary from hospital to hospital, but usually include the use of a stationary bike or treadmill. The most common method of exercise testing is the six-minute walk test, where the distance a patient can walk in six minutes is measured. Blood pressure, electrocardiogram, and oxygen saturation levels are monitored during exercise testing.

Esophogram: An X-ray examination of the esophagus (the tube that carries food to your stomach). This exam will help to determine if you suffer from GERD.

Echocardiogram (ECHO): A test that uses sound waves to create a picture of your heart, providing information about your heart function and screening for the presence of pulmonary hypertension.

treatment

THE CLINICAL COURSE OF PF IS HIGHLY VARIABLE and may be difficult to predict. As a result, strategies to treat PF are highly individualized, based upon the specific patient's medical history and other conditions. While there is no cure or FDA-approved treatments for IPF in the United States, and there are limited treatment options in the EU, Canada, and Asia, there are a variety of therapeutic options to help patients manage their condition and maintain their quality of life and activities of daily living. Typical standards of care may include prescription therapies, supplemental oxygen, pulmonary rehabilitation, lung transplantation, and/or referral for clinical trial participation. Lung transplantation remains the most viable course of treatment to extend the lives of those with IPF; this option should be discussed with your physician.

For some patients, depending upon their specific type of PF, medications may stabilize their disease and there may be a benefit to continuing usage. Further, some of these medications may be prescribed to manage symptoms when a patient has an acute exacerbation or period of worsening. Medications may be used alone or in combination.

It is important to note that patients with IPF taking or prescribed "triple combination therapy" with corticosteroids, azathioprine, and N-acetylcysteine (NAC) should discuss the risks and benefits of this combination therapy with their health care provider.[15]

The international IPF guidelines note that some therapeutic agents may provide a possible benefit for some patients. As with any medication for any condition, patients should discuss specific treatment options directly with their physician to determine the best approach for their care.

The following medications may be prescribed for the treatment of PF:

PHARMACOLOGIC TREATMENT OPTIONS

Corticosteroids (prednisone): Prednisone is used for suppressing the immune system and inflammation. It mimics the action of cortisol that is produced by the adrenal glands. Depending on the dose, prolonged therapy can cause the adrenal glands to stop producing cortisol. For this reason, when prednisone is discontinued, it may be necessary to gradually lower or taper the dose to allow time for the adrenal glands to recover. Since prednisone suppresses the immune system, it can potentially increase the frequency and severity of infections. Prednisone has many side effects, so individuals receiving prolonged treatment or higher doses need to be carefully monitored.

Cyclophosphamide (Cytoxan®): Cytoxan® is an anticancer drug frequently given in conjunction with prednisone or that may be given alone. While it is usually taken daily by mouth, in some instances it may also be administered intravenously.

Azathioprine (Imuran®): Imuran® is used to suppress the immune system and is commonly used to treat autoimmune diseases such as RA. It is also used to help prevent the body from rejecting organs following transplantation. Although there have been some successful reports in a small number of individuals, the effectiveness of Imuran® to treat IPF has not been confirmed in a randomized clinical trial to date.

N-acetylcysteine (NAC): NAC is a naturally occurring antioxidant. It can be taken orally and theoretically could prevent some of the oxidative injury that precedes an increase in fibroproliferation. A small, non-randomized study demonstrated some improvement in lung function in patients with IPF. There are a number of ongoing studies investigating the efficacy of NAC in combination with other drugs to treat IPF.

Pirfenidone (Esbriet®, Pirfenex®, Pirespa®): Pirfenidone is an anti-fibrotic and anti-inflammatory drug approved to treat mild-to-moderate IPF in the EU, Canada, and Asia. In other countries, including the US, pirfenidone is still undergoing clinical trials to investigate the efficacy of pirfenidone to treat IPF in order to meet regulatory requirements.

Supplemental Oxygen Therapy: As fibrosis inhibits an adequate transfer of oxygen into the bloodstream, some patients may require supplemental oxygen. This helps to reduce breathlessness, enabling the patient to be more active. Some patients may need oxygen therapy all the time while others may only need it during sleep and exercise. By testing the saturation level of oxygen in a patient's blood, a physician can determine if a patient requires supplemental oxygen.

If your doctor has prescribed supplemental oxygen, it is important to use it as prescribed. Many patients are fearful that they will become "addicted" to supplemental oxygen. It is important to recognize that supplemental oxygen is not addictive. The proper amount of oxygen in the bloodstream is necessary to maintain normal body functions. Low blood oxygen levels can lead to additional health problems.

It is important to note that medications utilized in the treatment of PF can vary by phenotype or cause of PF.

In addition to pharmacological treatments, patients with PF have non-drug options for treatment:

NON-PHARMACOLOGIC TREATMENT OPTIONS

Pulmonary Rehabilitation: Pulmonary rehabilitation includes conditioning; exercise training and breathing exercises; anxiety, stress, and depression management; nutritional counseling; education; and other components. The goal of pulmonary rehabilitation is to restore the patient's ability to function without extreme breathlessness. It has become the standard of care for people with chronic lung disease, and recent studies have demonstrated improvements in both exercise capacity and health-related quality of life in patients with IPF.[16] These programs offer a variety of services and can be inpatient, outpatient, or home/community based. The programs are multidisciplinary, meaning that the team includes nurses, respiratory therapists, physical therapists, social workers, dieticians, and others.

Lung Transplantation: IPF is now the leading indication for lung transplantation in many large transplant centers. Transplantation can improve both longevity and the quality of life in properly selected patients who have no other significant health problems. Previously, it was uncommon for individuals over the age of 65 to receive transplants. However, as surgical techniques and outcomes have improved more centers are performing transplants in individuals over age 65.

Until recently, early referrals were essential because of long pre-transplant wait times. Fortunately, with the new lung allocation system (LAS) used by the United Network for Organ Sharing (UNOS, www.unos.org) transplant candidates are evaluated based on the severity of their disease. As a result, wait times for transplantation have been dramatically reduced.

Transplantation is not without risk; patients should discuss all of the potential risks and benefits of lung transplantation with their physician.

TREATMENT OF ACUTE EXACERBATIONS AND COMORBIDITIES

Patients with PF can experience periods of worsening called acute exacerbations. High-dose prednisone is usually prescribed for these episodes.[2] Your doctor may provide prednisone or other treatments if you have an acute exacerbation.

Patients with PF frequently have other associated conditions, called comorbidities, as well. These can include pulmonary hypertension, GERD, obesity, emphysema, and obstructive sleep apnea. Your treatment plan will likely include both treatments that are directed at your PF and treatments for your comorbidities. Your care providers will help you determine how your comorbidities should be treated.

CLINICAL TRIALS

Since there are currently no FDA-approved therapies to treat IPF in the United States, and limited treatment options in the EU, Canada, and Asia, many patients choose to participate in clinical trials after consulting with their physician. New, experimental therapies are tested for their effectiveness through clinical trials.

The research community is aggressively investigating new treatments for all forms of PF. While the long-term goal of research is to prevent and cure the disease, present therapeutic approaches consist of attempts to slow disease progression and to extend the life expectancy of PF patients. While some studies are in advanced stages of development, others are in much earlier stages. There are a variety of clinical trials that are actively seeking the participation of patients. Some of the therapeutic approaches currently being studied include:

- Pulmonary vasodilators (such as sildenafil), which may aid in processing oxygen more efficiently.

- Anti-fibrotic therapies, which may slow or inhibit the production of scar tissue (fibrosis).
- Inhibitors of tumor angiogenesis, which block the signaling pathway of proteins shown to promote fibrotic proliferation.
- Inhibitors of "growth factor" proteins, which block proteins that can contribute to the formation of fibrosis.
- Genetic research to identify genes and genetic variants that may be associated with the development and progression of PF.
- Biomarker research looking for biologic molecules in the blood, tissue, or other body fluids that may predict the development of PF, rate of disease progression, or efficacy of a therapeutic intervention.

It is very important that patients discuss the possibility of participating in a clinical trial with their physician upon diagnosis. It is through clinical trials that a cure for the disease will be found. Please visit the research section of our website at www.pulmonaryfibrosis.org/research to learn more about clinical trials.

monitoring

CONTINUED MONITORING OF YOUR PF is a very important part of maintaining your health. Through monitoring, you and your care providers can determine how well you are responding to your treatment, whether your disease is stable, and what next steps should be taken. Regular interactions with your care providers will also help ensure that you receive the most current and best possible PF treatments.

Your clinical monitoring pattern will vary dependent upon your specific type of PF. Regardless of the underlying cause of your PF, continued monitoring is a vital component in your treatment. Talk with your provider about how frequently you should see them and what steps need to be taken to ensure that your disease is properly managed.

palliative and hospice care

THE MAIN GOAL OF BOTH PALLIATIVE AND HOSPICE CARE IS MAINTAINING PATIENT COMFORT. Palliative care does not specifically treat PF, but is care designed to improve the quality of life for patients with a chronic illness. Accordingly, it is appropriate for any patient who experiences discomfort due to PF.

PALLIATIVE CARE

Using a multidisciplinary approach, palliative care can involve physical, psychosocial, and spiritual factors in the treatment approach. Teams may include physicians, pharmacists, nurses, religious leaders, social workers, psychologists, and other health care professionals. In patients with PF, these teams focus on concrete goals including relief from pain or other distressing symptoms, spiritual care, development of support systems, and encouraging an active lifestyle.

HOSPICE CARE

Hospice care is a type of end-of-life care; it is intended to help people who are dying have peace, comfort, and dignity. It is generally reserved for patients who have less than six months to live. Patients in hospice care receive treatments to control pain and other symptoms to maintain comfort. Hospice care also provides support to families. Care may be provided at a hospice center, but can also be done in nursing facilities, hospitals, or at home.

lifestyle changes

THERE ARE A VARIETY OF THINGS that patients can do to maintain or improve their quality of life while living with PF. The National Institutes of Health (www.nih.gov) and the Mayo Clinic (www.mayoclinic.org) offer a variety of recommendations for patients, some of which we have referenced in this section.

Stay in Shape. The most damaging consequence of lung disease and its sensation of breathlessness is the development of an inactive lifestyle. For many patients, activities of daily living, like bathing and dressing, can create overwhelming fatigue. "Air hunger" can create panic attacks, and produce negative psychological effects. People with chronic respiratory problems sometimes limit their physical activities in an attempt to avoid shortness of breath. The lack of exercise works against you; inactivity weakens your muscles and they become less efficient. Deconditioning can make even the simplest daily activities more difficult. Regular exercise strengthens your muscles and makes them more resistant to fatigue. With practice and training you can learn to perform tasks in a more efficient manner. By being more efficient you need less oxygen for the same amount of work. The result is that you may find that you have more energy to accomplish daily tasks and that you are less short of breath. A formal rehabilitation program (pulmonary rehabilitation) is preferred because it allows for observation during exercise and it can be tailored to your specific needs.

Eat Well. A healthy diet includes a variety of fruits, vegetables, and whole grains. It also includes lean meats, poultry, fish, beans, and fat-free or low-fat dairy products. A healthy diet is low in saturated fat, trans fat, cholesterol, sodium (salt), and added sugar. Eating smaller, more frequent meals may

prevent stomach fullness that can make it harder to breathe. If you need help with your diet, ask your doctor to arrange for a dietician to work with you. A nutritionally rich diet that contains adequate calories is essential. A dietician can give you further guidelines for healthy eating.

Get Plenty of Rest. Getting at least eight hours of quality rest every night can boost your immune system and sense of well-being.

Stop Tobacco Use. Avoiding environmental irritants, like cigarette smoke, is a good way to prevent further damage to your lungs. If you are still smoking, the most important thing you can do is to stop. Due to the addictive nature of tobacco, this can be difficult. Seek the help of your physician to find a smoking cessation class or other beneficial methods to help you stop smoking. Secondhand smoke can be as harmful to you as if you were smoking yourself. Ask your family and friends to refrain from smoking around you.

Learn and Practice Relaxation Techniques. When you are physically and emotionally relaxed, you avoid excessive oxygen consumption caused by tension of overworked muscles. Additionally, learning relaxation techniques can help you manage the panic that often accompanies shortness of breath. Joining a support group and/or seeing a counselor can help you cope with your feelings and the anxiety and depression that are common in people with chronic breathing disorders; these feelings may aggravate the underlining disease. Many fear losing the ability to function and becoming dependent on others. The restriction on activity due to shortness of breath may lead to isolating oneself from family and friends, adding to the depression.

Join a Support Group. Just knowing that there is someone out there that knows how you feel is comforting. Share ideas, share fears, and share joys. A detailed listing of local and online support groups can be found at www.pulmonaryfibrosis.org/supportgroups.

Participate in Your Health Care. Remember you are part of a health care team that includes your doctors and nurses. They will be asking you a lot of questions. As a member of that team you have a responsibility to do your part. Be prepared to ask your own questions. Be a participant. Bring someone with you to each appointment and prepare a list of questions to be answered by your physician during your visit.

Help Others with PF. Consider participating in the Pulmonary Fibrosis Foundation's advocacy program. You may gain strength in knowing that you are helping future patients and researchers by advocating for the pulmonary fibrosis community.

Keep a Positive Attitude. Actively participating in the management of your disease is greatly enhanced by a positive attitude. A positive attitude can help you and your loved ones cope with your disease.

maintaining your care

YOU ARE THE CENTER OF YOUR TREATMENT. While your doctors, nurses, and other care providers will help you manage your disease, *you are your most important advocate*. People who take an active role in their own care do better over time. Here are a few steps you can take to make sure you maximize your care:

Speak up for yourself. If you have any concerns with your treatment or do not understand something about your disease, talk to your care providers. They want to make sure that you are able to maintain your health and will help you with these issues.

Be prepared for your visits. Ensure that you are able to see your care providers regularly. Have a list of any questions or concerns.

Ask questions of your care providers. This goes along in communicating with your providers and being prepared. You cannot help in the decision making process if you do not understand the factors involved.

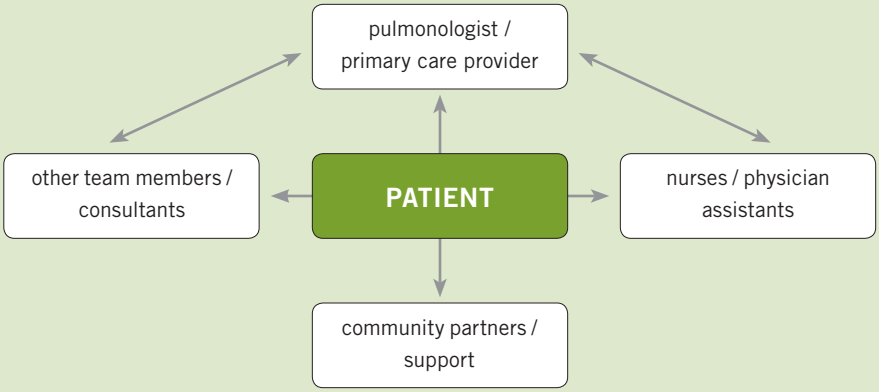
Take notes. You will likely get a lot of information during your health care visits and this can be overwhelming. Do not be afraid to take notes to help you remember important treatment issues later on.

Let your family and friends help. Emotional support is as important as other treatments. You can bring people who support you to your health appointments. Additionally, maintaining your health is a lot easier if you have support. This is especially true if you are trying to modify your lifestyle. Quitting smoking, exercising more, or changing your diet is difficult; let those who care about you help you accomplish your goals.

Contact advocacy and support groups. Reach out to community or national groups that help people with PF. It can be very useful to find out how other people manage their disease. They may give you invaluable tips or tools that make it easier for you to manage your disease. You can also pass your tips on to others and help support the community.

Stay informed. Keep learning about PF. The more you know, the better you will be able to manage your disease and recognize when you need to access health services. Be warned that there is misinformation on the internet; make sure you access reliable sources of information, such as advocacy groups and respected scientific and medical sources.

YOUR CARE TEAM



questions to ask your health care providers

YOUR CARE PROVIDERS ARE PARTNERS in your PF treatment. It is important that you have a firm understanding of your disease and how you should care for it. Ask your providers about anything relating to your treatment that you do not fully understand. Here are a few questions that may help you manage your care:

How will PF impact what I can and cannot do?

You probably already know the limits of what you can and cannot do in your everyday routine. There may be activities that you do not regularly engage in which may be impacted by your PF. This can include traveling by air or visiting high-altitude places. Your care providers can help you identify what activities may present some challenges.

What should I do and whom should I contact if I have any problems with my PF?

Having an action plan in place in case of problems is a must for anyone with a chronic disease. Ask about where you should go, who you should call, and what you should do during nights and weekends if you have a problem with your PF. You should also know who to contact if you have any questions and concerns about your current care, including your treatment.

When is the right time to start or switch treatment?

The decision to start therapy depends on your health and desires. It should be made in conjunction with your health care providers and those who will help support you, such as family and friends. Once you have started therapy, you may need to change it as your disease and needs evolve. Of course, when your treatment is not working well it is time to discuss other treatment options. However, even when treatment is working there may be better options for your needs.

Asking about what options are available is a good way to find out what changes are happening in the treatment of PF and helps make sure that you get the best treatment for both your disease and lifestyle.

Will my treatment interfere with other medications I am taking?

It is very important that your care providers know all of the medications that you take regularly—both prescribed and over-the-counter—as well as any vitamins or other nutritional supplements. Asking your providers about drug-drug interactions helps ensure that all of your medications are reviewed for potentially dangerous interactions.

Are there resources that can help me lead a healthier lifestyle?

Improving your lifestyle is a critical component of improving quality of life. However, changing things like how much you exercise, your diet, and smoking habits can be very difficult. Care providers may be able to direct you to resources that can help you modify your lifestyle and adopt healthier behaviors. These can include support groups, dieticians, and personal trainers. Showing an interest in adopting healthier behaviors is the first step in accomplishing these changes.

Is a clinical trial right for me?

Clinical trials are a potential resource for patients who may not have many good treatment options. However, whether you should enroll in a clinical trial depends on many factors, including what trials are available in your area, whether you are motivated to participate, and your current condition. If you are interested in participating in a clinical trial, talk with your care providers about options; they will help you determine if a clinical trial is right for you.

about the pulmonary fibrosis foundation

THE MISSION OF THE PULMONARY FIBROSIS FOUNDATION (PFF) is to help find a cure for idiopathic pulmonary fibrosis (IPF), advocate for the pulmonary fibrosis (PF) community, promote disease awareness, and provide a compassionate environment for patients and their families.

The PFF's strategic plan includes initiatives to:

- Increase funding for PF research through independent foundation grants, and partnership grants with the American Thoracic Society, the American College of Chest Physicians, and the NIH.
- Facilitate collaboration between the academic research community and the bio-pharma industry.
- Establish a Pulmonary Fibrosis Patient Registry and Clinical Care Network.
- Foster interaction and innovation among physicians, researchers, allied health professionals, patients, and caregivers at our biennial international conference *PFF Summit: From Bench to Bedside.*
- Expand our support group network to include the international PF community, assist in the development of local support groups, and improve access to the PFF online support groups.
- Implement new patient education and disease awareness programs utilizing webinars, online support services, and social media platforms.
- Support the needs of our constituents through legislative advocacy.
- Increase disease awareness though education, traditional media, social media, and community events.

Our staff is always available to discuss your individual needs. If you know of a patient or family that could use our help, please feel free to share our contact information.

Pulmonary Fibrosis Foundation
230 East Ohio Street, Suite 304
Chicago, Illinois 60611-3201

888.733.6741 Phone info@pulmonaryfibrosis.org
866.587.9158 Fax www.pulmonaryfibrosis.org

JOIN THE PFF COMMUNITY

We need your help! Joining the Pulmonary Fibrosis Foundation is free of charge and will help you better connect with the pulmonary fibrosis community as we strive to cure this devastating disease. Benefits include:

- Invitations to PFF sponsored educational events including webinars
- Participation in our online communities and support groups
- Our monthly e-newsletter and biannual *Breathe Bulletin*
- Email updates important to the PF community
  - – Support group announcements
  - – Clinical trial announcements
  - – Fundraising announcements and invitations
- Participation in PFF advocacy efforts

How can you invest in helping to find a cure for IPF?

- Make a gift of cash
- Make a gift of marketable securities
- Purchase a PFF "Breathe" bracelet and other PFF products
- Name the PFF in your family will and bequests
- Establish a charitable gift annuity for the benefit of the PFF
- Become a volunteer

Call 888.733.6741 or visit www.pulmonaryfibrosis.org to make a gift or join the PFF community.

glossary

Acute exacerbation: An episode of rapid decline or the emergence of symptoms.

Alveoli: Tiny air sacs in the lungs where carbon dioxide leaves the bloodstream and oxygen enters the bloodstream.

Bronchoscope: A tool used for inspecting the inside of the lungs.

Comorbidity: A disease or other issue that occurs simultaneously with PF.

Diffuse parencyhmal lung diseases (DPLD): Another name for interstitial lung diseases.

Diffusion capacity (DLCO): A measure of the ability of oxygen to diffuse into the bloodstream.

Dyspnea: Difficulty breathing or shortness of breath.

Fibroproliferation: Of or relating to the growth of fibroblasts, one of the basic connective tissue cells.

Fibrosis: An increase in fibrous scar tissue.

Forced expiratory volume (FEV1): The amount of air you can blow out in one second. Measured by spirometry.

Forced vital capacity (FVC): How much air you can blow out of your lungs. Measured by spirometry.

Gastroesophageal reflux disease (GERD): A regurgitation of stomach acids into the esophagus and throat, causing heartburn, acid indigestion, and possibly injury to the lining of the esophagus. Also called acid reflux disease.

Hospice care: Palliative care for patients at end-of-life.

Idiopathic: Of unknown cause.

Idiopathic interstitial pneumonias (IIP): A type of interstitial lung disease. IPF is a type of IIP.

Interstitial lung diseases (ILD): A broad category of over 200 lung diseases that affect the lung interstitium.

Interstitium: The space around the alveoli.

Palliative care: Non-curative therapy that treats symptoms and focuses on improving quality of life. It can be received at the same time as curative therapy.

Pathologist: A physician specializing in disease-associated changes in tissue and organs. Pathologists aid in medical diagnosis.

Pulmonary: Relating to the lungs.

Pulmonary hypertension: Abnormal high blood pressure in the lung arteries.

Pulmonologist: A physician specializing in the lungs.

Radiologist: A physician specializing in using radiology tests (e.g., X-rays) to diagnose illness.

Rheumatologist: A physician specializing in rheumatic diseases, which may include arthritis, autoimmune diseases, and joint diseases.

Spirometry: A test that measures the amount of air inhaled and exhaled over time.

Usual interstitial pneumonia (UIP): A specific abnormal radiologic or pathologic pattern.

references

1 National Heart Lung and Blood Institute. "What is idiopathic pulmonary fibrosis?" Available at: www.nhlbi.nih.gov/health/health-topics/topics/ipf/. Accessed June 29, 2012.

2 Raghu G, Collard HR, Egan JJ, et al. "An official ATS/ERS/JRS/ALAT statement: idiopathic pulmonary fibrosis: evidence-based guidelines for diagnosis and management." *Am J Respir Crit Care Med* 2011;183:788–824.

3 King TE Jr. "Clinical advances in the diagnosis and therapy of the interstitial lung diseases." *Am J Respir Crit Care Med* 2005;172:268–279.

4 Raghu G, Weycker D, Edelsberg J, Bradford WZ, Oster G. "Incidence and prevalence of idiopathic pulmonary fibrosis." *Am J Respir Crit Care Med* 2006;174:810–816.

5 Navaratnum V, Fleming KM, West J, et al. "The rising incidence of idiopathic pulmonary fibrosis in the UK." *Thorax* 2011;66:462–467.

6 Olson AL, Swigris JJ, Lezotte DC, Norris JM, Wilson CG, Brown KK. "Mortality from pulmonary fibrosis increased in the United States from 1992–2003." *Am J Respir Crit Care Med* 2007;176:277–284.

7 Collard HR, Tino G, Noble PW, et al. "Patient experiences with pulmonary fibrosis." *Respir Med* 2007;101:1350–1354.

8 Mayberry JP, Primack SL, Muller NL. "Thoracic manifestations of systemic autoimmune diseases: radiographic and high-resolution CT findings." *RadioGraphics* 2000;20:1623–1635.

9 Nogee LM, Dunbar AE 3rd, Wert S, Askin F, Hamvas A, Whitsett JA. "Mutations in the surfactant protein C gene associated with interstitial lung disease." *Chest* 2002;121:20S–21S.

10 Rosas IO, Richards TJ, Konishi K, et al. "MMP1 and MMP7 as potential peripheral blood biomarkers in idiopathic pulmonary fibrosis." *PLoS Med* 2008;5:e93.

11 Armanios MY, Chen JJ, Cogan JD, et al. "Telomerase mutations in families with idiopathic pulmonary fibrosis." *N Engl J Med* 2007;356:1317–1326.

12 Alder JK, Chen JJ, Lancaster L, et al. "Short telomeres are a risk factor for idiopathic pulmonary fibrosis." *Proc Natl Acad Sci U S A* 2008;105:13051–13056.

13 Seibold MA, Wise AL, Speer MC, et al. "A common MUC5B promoter polymorphism and pulmonary fibrosis." *N Engl J Med* 2011;364:1503–1512.

14 American Thoracic Society. "Idiopathic pulmonary fibrosis: diagnosis and treatment (international consensus statement)." *Am J Respir Crit Care Med* 2000;161:646–664.

15 Wells AU, Behr J, Costabel U, Cottin V, Poletti V. "Triple therapy in idiopathic pulmonary fibrosis: an alarming press release." *Eur Respir J* 2012;39:805–806.

16 Nishiyama O, Kondoh Y, Kimura T, et al. "Effects of pulmonary rehabilitation in patients with IPF." *Respirology* 2008;13:394–399.

The Pulmonary Fibrosis Foundation is thankful to the following for reviewing this guide:

GREGORY P. COSGROVE, MD
National Jewish Health
University of Colorado
Denver, Colorado
PFF Medical Advisory Board Member

Boehringer Ingelheim

Translations of the *Pulmonary Fibrosis Patient Information Guide* were made possible by PLATINUM sponsor Boehringer Ingelheim and BRONZE sponsor InterMune.

PULMONARY FIBROSIS FOUNDATION

230 East Ohio Street, Suite 304

Chicago, Illinois 60611-3201

888.733.6741 phone

866.587.9158 fax

www.pulmonaryfibrosis.org